Stewart-Bluefarb Acroangiodermatitis in a Case of Parkes-Weber Syndrome

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Abstract
Stewart-Bluefarb acroangiodermatitis is the occurrence of pseudo-kaposiform changes with an underlying arterio-venous malformation. Parkes-Weber syndrome is a triad of arterio-venous malformation with varicose veins with bony and soft-tissue hypertrophy involving the extremity. A 13 year-old-female presented with ulcerated growth with bleeding episodes on right leg since past four years. A reddish discoloration over right leg was noticed at four years of age which remained unchanged until nine years of age, after which it showed rapid increase in size along with disproportionate increase in length and girth of right lower limb. Examination revealed hyperpigmented soft verrucous plaque over the right lower one-third of leg, along with presence of ulcers and dilated tortuous veins in the right lower leg with thrill and bruit over the right popliteal and inguinal region. A differential diagnosis of verrucous hemangioma and arterio-venous malformation with pseudo-kaposiform change was considered. Skin biopsy showed multiple fenestrated capillaries with perivascular lymphocyte infiltrate suggestive of capillary malformations. X-ray showed osteolytic defect in right tibia. Duplex ultrasound and magnetic resonance imaging of right leg showed multiple fast flowing small and medium sized arterio-venous malformations of small to moderate size. Thus, on the basis of clinical and radiological features, we made a diagnosis of Stewart-Bluefarb type of acroangiodermatitis with Parkes-Weber syndrome. She was managed conservatively with compression stockings.

Key Words: Parkes-Weber syndrome, Stewart-Bluefarb acroangiodermatitis, Stewart-Bluefarb syndrome

Introduction
Stewart-Bluefarb syndrome[1] consists of a pigmented purpuric eruption over the skin of lower limbs (acroangiodermatitis), clinically resembling Kaposi sarcoma (pseudo-kaposi sarcoma) overlying multiple arterio-venous malformations.[2] Parkes-Weber syndrome consists of arterio-venous malformations, varicose veins with limb hypertrophy. It commonly involves the lower limbs, lymphatic anomalies and lymphedema can be present.[3] In childhood usually areas of hypervascularity are seen radiographically in childhood, which forms arteriovenous fistulae at puberty or after trauma and can lead to systemic complications of cardiac enlargement and high cardiac output failure.

Case Report
A 13-year-old girl presented to us with a massive bleeding episode from an ulcer present on her right lower extremity. On enquiry she gave history of a red raised lesion over the right ankle area since she was four years of age. Since nine years of age she noticed increase in size of the lesion associated with disproportionate increase in dimensions of her right lower limb. There was no significant history of trauma. Medical, surgical and family history was non-contributory. Examination revealed hyper pigmented soft compressible plaques on lower one-third of right leg of approximately 15 × 10 cm in size. Few ulcers were present over the plaque with surrounding skin showing stasis changes [Figures 1 and 2]. Multiple dilated tortuous veins with prominent pulsations, audible thrill and palpable bruit were present on the involved limb. A limb-length discrepancy of 3 cm and limb-girth discrepancy of 7 cm was measured [Figure 3]. Routine hemogram, serum biochemistry and serology were within normal limits except for mild anemia. Culture studies from the ulcer showed growth of pseudomonas aeruginosa which was treated adequately with appropriate antibiotics. Skin biopsy showed proliferation of thick walled capillaries, erythrocyte extravasation, and fibroblastic proliferation with absence of promontory sign suggestive of pseudo-Kaposi sarcoma [Figure 4]. X-ray of right leg showed osteolytic defect in right upper tibia. Two dimensional-echocardiography revealed mild pulmonary hypertension. Arterial-venous Doppler showed multiple fast flowing arterio-venous malformations in the right lower limb which was confirmed by contrast enhanced magnetic resonance angiography and peripheral angiography [Figure 5]. Thus on the basis of clinical (limb hypertrophy, dilated tortuous vessels), histopathological (pseudo-Kaposi sarcoma) and
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radiological (fast flowing A-V malformations), a diagnosis of Stewart-Bluefarb type of acroangiodermatitis in a case of Parkes-Weber syndrome was made.

Discussion


Pseudo- Kaposi Sarcoma (acroangiodermatitis): This is reactive angiodyplasia of cutaneous blood vessels secondary to vascular causes. Its variants[5] are:

- Stewart-Bluefarb type: With underlying A-V malformation.
- Mali type: Stasis dermatitis with chronic venous insufficiency.
- Dermite ocre of Favre (gravity purpura): Venous varicosities with first pregnancy.
- Angiodermatitis over shunt for hemodialysis in chronic renal failure.

Figure 1: Hyperpigmented soft compressible plaques over lower 1/3 of right leg

Figure 2: Presence of ulcers over the plaque with stasis changes

Figure 3: Limb girth and length discrepancy with tortuous veins on right leg

Figure 4: Skin biopsy revealed proliferation of thick walled capillaries, red blood cell extravasation, and fibroblast proliferation suggestive of pseudo-Kaposi’s sarcoma (H and E, ×10)

Figure 5: Magnetic resonance imaging angiography revealed multiple dilated tortuous vessels on the right limb
The term “Stewart-Bluefarb syndrome” was coined in 1967 Bluefarb,[6] Adams and Stewart[7] where the cause of underlying malformations could be congenital, traumatic or iatrogenic.

The proposed pathogenesis is due to defective oxygen and carbon dioxide perfusion which leads to secretion of vascular endothelial growth factors which causes proliferation of endothelial cells and fibroblasts, leading to hypertrophy and dermatitis changes.[9]

The clinical features consist of soft compressible painful purple papules, nodules and plaques which may ulcerate. The clinical features of underlying A-V malformation/fistula includes dilated tortuous veins with palpable thrill and audible bruit on auscultation, enlargement of limb and increased local warmth. Complications of ulcers, secondary infections, torrential bleeding episodes, osteolytic bone changes, cardiac failure and pulmonary hypertension may be seen. Histopathology shows proliferation of thick walled capillaries with red blood cell extravasation, hemosiderin deposition and fibroblast proliferations.

Management consists of conservative modalities in the form of bed rest, limb elevation, compression bandages and symptomatic treatment of secondary complications. Surgical treatment consists of ultrasound guided sclerotherapy, selective embolisation, microvascular surgery (skeletonisation) and rarely amputation may be required. The management varies according to size, number and location of A-V Malformations. Repeated embolization of the feeding vessels under angiographic control is most effective.[1]

In our case, Parkes-Weber syndrome was the primary condition with multiple A-V malformations leading to the secondary complication of acroangiogdermatitis of Stewart-Bluefarb type. Onset of puberty in our patient had lead to proliferation of the A-V malformations thereby the plaques increased in size when patient was 9 years of age. Conservative line of management in the form of oral antibiotics, local dressings and compression stockings was given due to multiple malformations not being amenable to vascular surgery.

Conclusion

Management of acroangiogdermatitis consists of treating the underlying cause. Management of A-V malformation varies according to size, number and location of A-V Malformations. Puberty causes a proliferation of the A-V malformations. Sclerotherapy if used early can prevent secondary skin changes.

Table 1: Differences between Klippel-Trenaunay syndrome and Parkes-Weber syndrome

<table>
<thead>
<tr>
<th>Klippel-Trenaunay Syndrome</th>
<th>Parkes-Weber Syndrome</th>
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<tbody>
<tr>
<td>Features</td>
<td></td>
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<tr>
<td>Triad of port wine stain</td>
<td>Varicose veins+Limb</td>
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<tr>
<td>(capillary malformations)</td>
<td>Hypertrophy+A-V</td>
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<tr>
<td>+ Varicose veins+Limb</td>
<td>Malformation*</td>
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<tr>
<td>Hypertrophy</td>
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<tr>
<td>Genetics</td>
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<td>AGGF1 mutation</td>
<td>RASA1 mutation</td>
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<td>Abnormalities</td>
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<tr>
<td>Mesodermal</td>
<td>Ectodermal+Mesodermal</td>
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<tr>
<td>Type of flow</td>
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<td>Slow</td>
<td>Fast</td>
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<td>Prognosis</td>
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<tr>
<td>Favourable</td>
<td>Severe and Fatal</td>
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a: Differentiating feature between the two

References


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